



SHORT REPORT

Phlegmasia Cerulea Dolens of the Lower Extremities Secondary to Thrombosis of an Inferior Vena Caval Aneurysm

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Abstract We present the clinical case of a 34-year-old male with an aneurysm of the inferior vena cava in which thrombosis led to a picture of bilateral phlegmasia cerulea dolens of the lower extremities. A clear and precise diagnosis was achieved by angio CT which allowed initial conservative management with good immediate and short term outcome.

To our knowledge only 23 cases of aneurysm of the inferior vena cava have been reported in the literature worldwide making it difficult to establish a diagnostic and therapeutic algorithm for these cases. Nine cases were presented with thrombosis and in the other 14 the diagnosis was incidental. They mimic a pararenal mass requiring differential diagnosis with retroperitoneal neoplasms.

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Introduction

We present a patient with a thrombosed inferior caval aneurysm presenting with phlegmasia cerulea dolens. The clinical manifestations of thrombosis of the inferior vena cava (IVC) vary greatly, ranging from the absence of

symptoms to phlegmasia cerulea dolens. Aneurysms of the vena cava are extremely rare. To our knowledge, only 23 cases have been described in the literature, 9 presented with thrombosis (one of them with a fatal pulmonary embolism, and none with phlegmasia cerulea dolens) and the other 14 with incidental diagnosis. The mode of presentation did not appear to vary with either age or gender.

Case Report

A 34-year-old male had no known medication allergies and with bronchial asthma treated with salbutamol inhaler on

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demand. He attended the emergency ward of our hospital with a picture of sudden 4-hour onset of pain in the lower extremities and inability to walk. The lower extremities were swollen and tense with cyanosis up to inguinal region, poor differentiation of the muscular compartments, and pale, cold feet. The femoral and popliteal pulses were palpable bilaterally with absence of pedal and posterior tibial pulse in both extremities.

Following the initial evaluation and probable diagnosis of massive thrombosis of the IVC a computerized axial angiotomography of the abdomen was performed (Figs. 1 and 2). This showed hypoplasia of the suprarenal IVC in the intrahepatic segment, thrombosis of the left renal vein and a sacular aneurysm of the thrombosed IVC. The thrombosis was found to extend distally to the femoral veins involving the iliac region.

On achieving the definitive diagnosis of massive thrombosis of the IVC, the patient was transferred to the intensive care unit (ICU) for monitoring. He was placed in the Trendelenburg position and systemic anticoagulation was started. Distal pulses were recovered, with improvement in the perfusion of the feet. Following 72 hours of observation in the ICU the patient was transferred to the department of vascular surgery where oral anticoagulation was administered with a coumarin derivative.

After 10 days in hospital the patient was able to walk and was discharged with adjuvant compression therapy.

Discussion

In 1973 Oh KS *et al.* published the first case of aneurysm of the IVC. In 1993, Gradman WS *et al.* described the thirteenth case and reviewed the 12 previous cases to establish a classification of this rare entity.¹ They divided the aneurysms of the IVC into 4 types based on the localization of the aneurysm and the association of other venous abnormalities.

Four etiologic theories have been suggested. The first is congenital due to an alteration during embryonic development.² The second is continuous hypertension, secondary to obstruction in the venous drainage, which would eventually produce weakness of the vessel wall leading to venous dilatation.² Traumatic origin has been described (trauma which weakens the venous wall).² Last, an arteriovenous fistula may be responsible for the aneurysm.³ Our case possibly relates to the second etiology. However, the true etiology remains unknown.

It is difficult to establish a diagnostic algorithm with only 23 different cases having been reported. However,

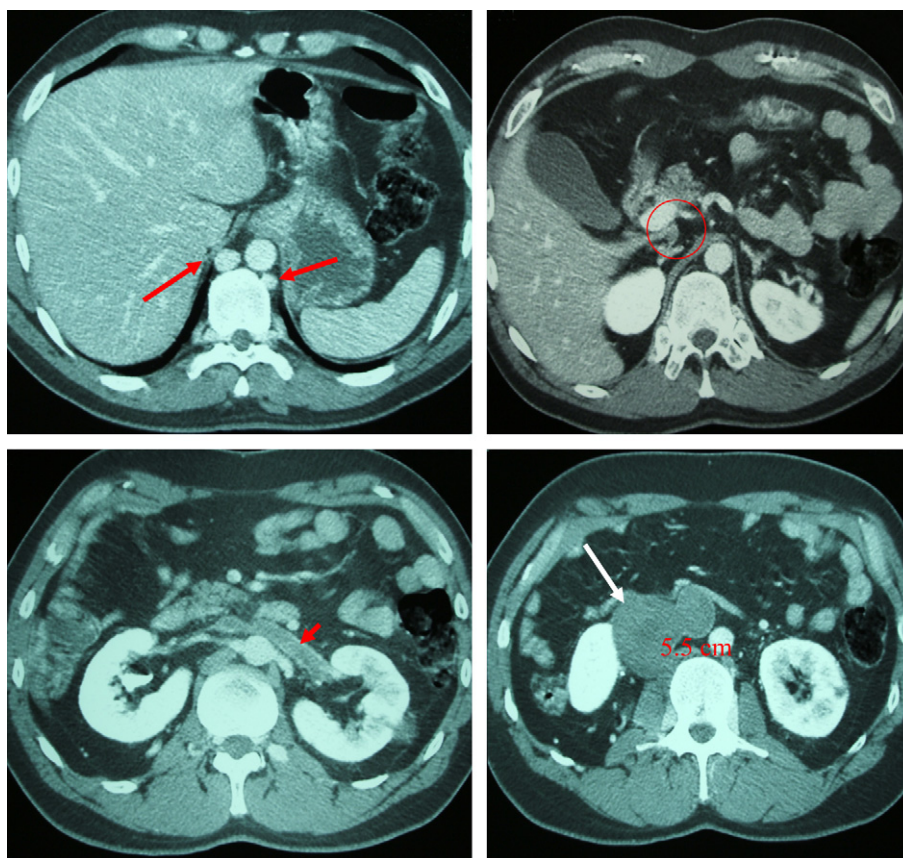


Figure 1 Suprarenal axial views. Hypertrophy of the azygos and hemiazygos systems (red arrows). Hypoplasia of the inferior vena cava in the intrahepatic segment (diameter 0.7 cm) (red circle). Infrarenal axial views. Observe the thrombosis of the left renal vein (short arrow). Right pararenal mass of homogeneous density with the thrombosed inferior vena cava of 5.5 cm of maximum diameter with cranial-caudal extension from the renal hilum to beyond the inferior renal poles (white arrow). The diameter of the normal inferior vena cava was 2 cm.

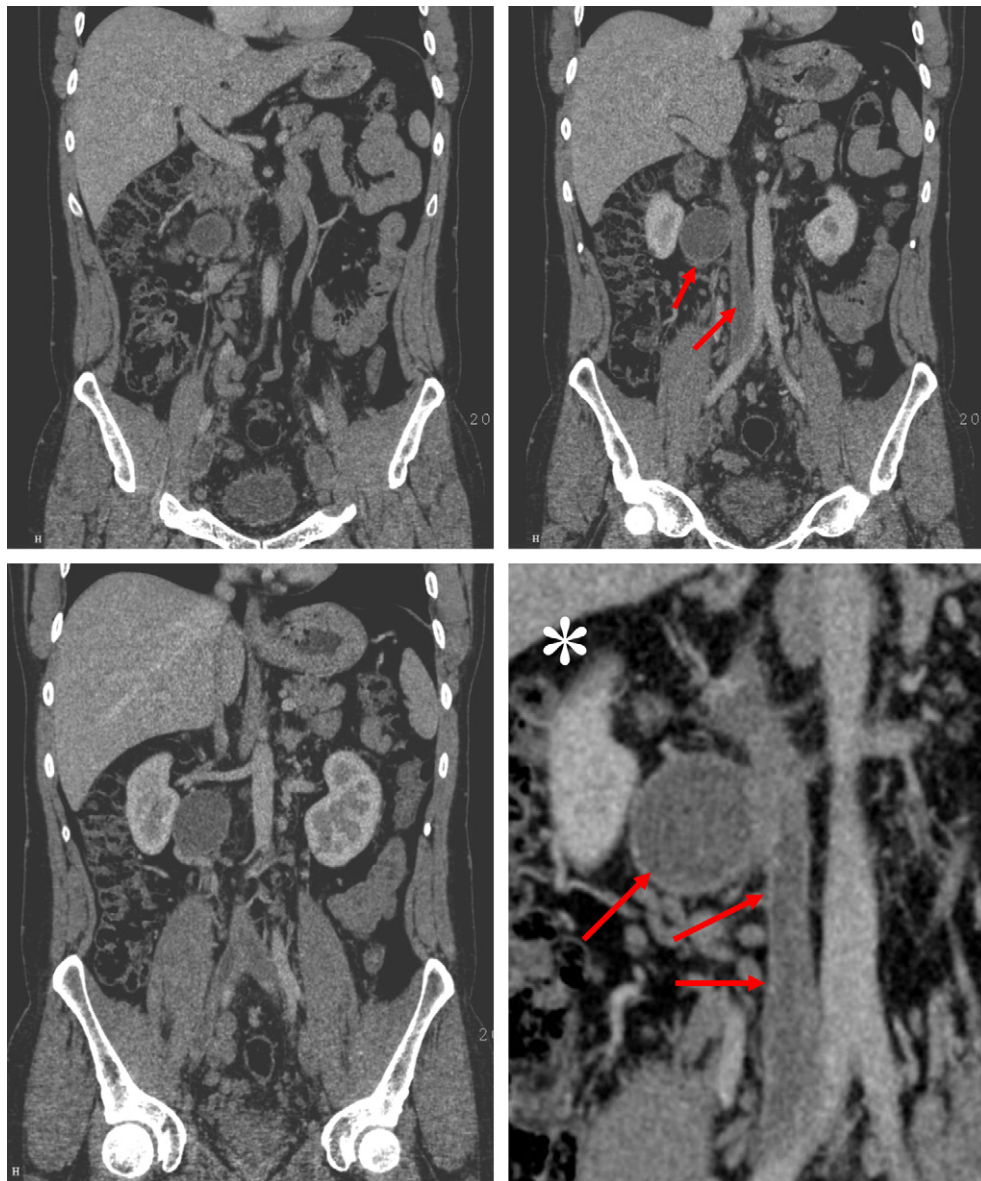


Figure 2 Coronal biplane reconstructions. The wall of the inferior vena cava may be seen continuing with the wall of the aneurysm (red arrows). *Rectangle area magnified.

the CT-angiography and/or the MR-angiography may be considered as the gold standard for achieving the definitive diagnosis, providing that there is no thrombosis of the aneurysm of the IVC.^{1,4} When the aneurysm presents with thrombosis angiographic imaging is unable to differentiate this entity from a retroperitoneal tumor. Positron emission tomography (PET) may clarify the nature of the mass.² Laparotomy with exploratory biopsy may be necessary in cases in where the benign nature cannot be established by imaging.^{1,4} In our case the tomographic imaging was definitive (Fig 2).

Biopsy of the aneurysm would show the three tunics of the vein to be normal but with different characteristics. The external and muscular tunica would be thinned and the internal tunica fragmented. Different membranous bands would cover the interior of the aneurysm.²

With regard to therapeutic management, several options have been used. Anticoagulation and observation are implemented as conservative treatment for incidental diagnosis of a non-thrombosed aneurysm of the IVC.² Laparotomy with biopsy of the aneurysm and vascular reconstruction are used in cases of non-thrombosis in patients with low surgical risk.³ Laparotomy with biopsy and proximal and distal ligation is performed for aneurysms with thrombosis either when a clear diagnosis cannot be achieved with imaging techniques or in the presence of large cranial-caudal extension of the thrombosis not allowing vascular reconstruction.^{2,4} Finally, local intravenous fibrinolysis with urokinase may be undertaken by venous access, followed by conservative follow up.⁵ The four therapeutic approaches mentioned above are those which have been described in the literature.

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